Acute coronary syndrome in a patient with multiple coronary artery fistulas draining from the left anterior descending artery into the left ventricle – 2-year follow-up

Ostry zespół wieńcowy u chorego z mnogimi przetokami gałęzi międzykomorowej przedniej do lewej komory serca – obserwacja 2-letnia

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Abstract

Coronary artery fistula (CAF) is an abnormal connection between a coronary artery and a chamber of the heart (most often the right ventricle) or a large vessel. Most fistulas remain asymptomatic and are discovered accidentally during coronary artery diagnostics performed for other reasons. It is assumed that clinical symptoms and chest pain or exertional dyspnoea are caused by a steal phenomenon. In the case of coronary artery fistulas therapeutic management depends on clinical manifestation and the significance of haemodynamic consequences caused by the fistula. It should be noted that current guidelines of treatment are based on small retrospective studies.

We present a case of a 45-year-old patient with CAF of a rare location draining from the left anterior descending artery into the left ventricle and accompanied by a myocardial bridge narrowing the lumen of this artery. The patient presented with symptoms of acute coronary syndrome.

Key words: coronary artery fistula, acute coronary syndrome

Streszczenie

Przetoka tętnicy wieńcowej (CAF) jest nieprawidłowym połączeniem między tętnicą wieńcową a jamą serca (najczęściej prawą komorą) bądź dużym naczyniem. Większość przetok pozostaje bezobjawowa a rozpoznawane są przeważnie przy okazji diagnostyki naczyń wieńcowych przeprowadzanej z innych przyczyn. Uważa się, że objawy kliniczne takie jak ból w klatce piersiowej czy duszność wysiłkowa są spowodowane efektem podkradania. Postępowanie terapeutyczne w przypadku przetok tętnic wieńcowych jest zależne od manifestacji klinicznej i istotności następstw hemodynamicznych powodowanych przez przetokę. Należy dodać, że istniejące wytyczne postępowania zostały opracowane na podstawie niewielkich badań retrospektywnych.

Przedstawiamy przypadek 45-letniego chorego z CAF o rzadkiej lokalizacji, łączącej gałąź międzykomorową przednią lewej tętnicy wieńcowej z lewą komorą, z towarzyszącym mostem mięśniowym zawężającym światło tego naczynia. Pacjent prezentował objawy ostrego zespołu wieńcowego.

Słowa kluczowe: przetoka tętnicy wieńcowej, ostry zespół wieńcowy

Introduction

Coronary artery fistula (CAF) is a rare malformation caused by an abnormal connection between a coronary artery and neighbouring structures, most often the right ventricle (approx. 40% of cases). Fistulas drain to (in decreasing frequency of occurrence) the right atrium, pulmonary artery and coronary sinus. In only 2% of cases fistulas connect coronary arteries with the left ventricle.

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In 55% of cases CAF originate from the right coronary artery (RCA), in 35% from the left coronary artery (LCA) and in 5% of cases from both coronary arteries at the same time [1].

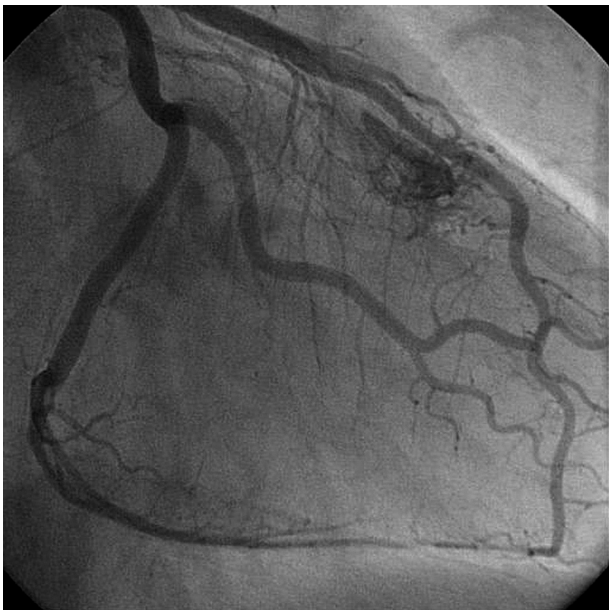

Fig. 1. Angiography of the left coronary artery. Multiple coronary artery fistulas draining from perforating branches of the left anterior descending artery to the left ventricle

Ryc. 1. *Angiografia lewej tętnicy wieńcowej. Widoczne mnogie przetoki pomiędzy gałęziami przeszywającymi odchodzącymi od LAD a lewą komorą*

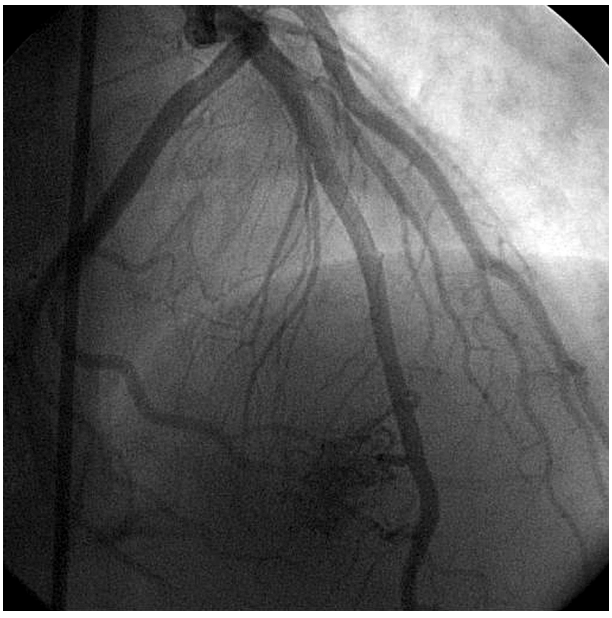

Fig. 2. Coronary artery fistulas between perforating branches of the left anterior descending artery and the left ventricle

Ryc. 2. *Przetoki pomiędzy gałęziami przeszywającymi odchodzącymi od LAD a lewą komorą*

The prevalence of this abnormality according to various sources is believed to reach 0.1-0.87% of all coronary angiographies and 0.001-0.002% in the general population [2-6]. We present a case of a patient with a coronary artery fistula draining from the left anterior descending artery (LAD) into the left ventricle.

Case report

A 45-year-old man, previously untreated, a smoker (around 20 cigarettes per day for 25 years), with a history of arterial hypertension and hyperlipidaemia, was admitted to our centre in the second hour of a strong resting chest pain which appeared after alcohol consumption. Similar symptoms but of a smaller intensity had occurred spontaneously in this patient on exertion for several years. ECG on admission showed regular sinus rhythm of 70 bpm with 1.5 mm high ST-segment elevation in leads V_2-V_4. Blood pressure was 140/90 mmHg. There was a silent systolic bruit on heart auscultation. Transthoracic echocardiography did not disclose any regional wall motion abnormalities, no enlargement of the ventricles (left ventricle = 51 mm at end-diastole), no pathological left-to-right shunts or pulmonary hypertension. There was only mild mitral regurgitation present. Left ventricular ejection fraction was 64%. The chest X-ray was normal. Combined antiplatelet treatment with loading doses of 300 mg of aspirin and 600 mg of clopidogrel, unfractionated heparin (5000 IU), ramipril, atorvastatin and proton pump inhibitor (pantoprazole) were used. Immediate coronary angiography

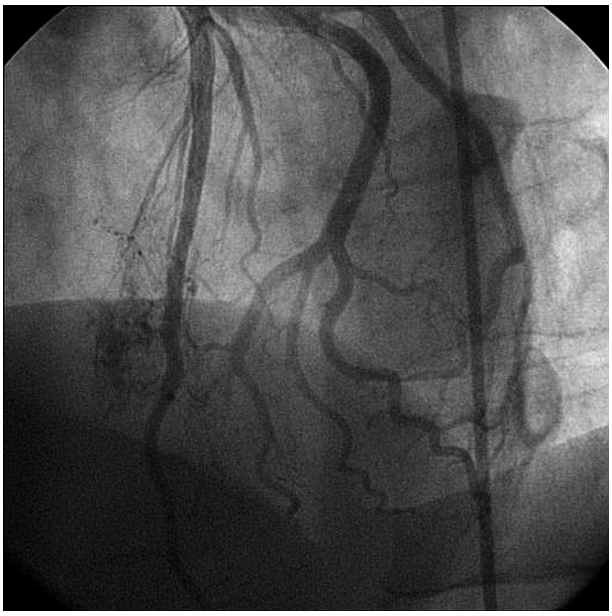

Fig. 3. Left coronary artery. Multiple CAF draining from perforating branches of the left anterior descending artery to the left ventricle

Ryc. 3. *Lewa tętnica wieńcowa. Liczne przetoki pomiędzy gałęziami przeszywającymi odchodzącymi od LAD a lewą komorą*

showed a myocardial bridge over the LAD narrowing the artery in systole by 40-50% and no changes in other coronary arteries. Another finding was fistulas between three perforating branches arising from the LAD and the left ventricle (fig. 1-3). Myocardial necrosis markers remained unelevated in repeated samples, ECG changes normalized and there were no recurrences of angina. Exercise test was normal and there were no arrhythmias on ambulatory Holter ECG. The myocardial bridge and alcohol consumption (increasing the steal phenomenon) were considered as the cause of acute ischaemia. The patient was diagnosed with unstable angina (acute coronary syndrome with ST-segment elevation on admission) and fistulas draining from the left anterior descending artery into the left ventricle with a coexisting myocardial bridge. Despite symptoms of acute coronary syndrome, in view of the lack of haemodynamic significance of the fistula (no volume overload or left ventricular hypertrophy), good exercise tolerance and no consent from the patient for surgical management, together with a lack of studies univocally confirming the efficacy of this method, a conservative treatment was chosen with periodic monitoring using imaging techniques. The patient was discharged home in good general condition with the following instructions: complete smoking cessation, limitation of alcohol consumption, systematic cardiological follow-up and 12 months of combined dual antiplatelet therapy accompanied by pantoprazole. Life-long treatment with aspirin, ramipril and atorvastatin were recommended.

A follow-up visit was conducted 13 months after the acute coronary syndrome. The patient did not report any recurrences of symptoms since hospitalization. He had not stopped cigarette smoking despite the recommendation. A control exercise test was performed, but there was no chest pain on exertion and ECG remained normal. The test was ended at peak heart rate for sex and age. A 24-hour ambulatory Holter ECG did not show any arrhythmias.

Currently, 24 months after the incident the patient remains asymptomatic, has good exercise tolerance, ventricles are not enlarged and there are no signs of left ventricular overload.

Discussion

Coronary artery fistula was observed for the first time at autopsy and described by Krause in 1865 [7]. In 50% of cases coronary artery fistulas are congenital malformations. Aetiopathogenesis of congenital fistulas communicating with the left ventricle is related to the presence of a network of capillaries draining to the lumen of the left ventricle in the early stages of embryogenesis. During myocardial growth the lumen of the capillaries becomes occluded and the capillaries eventually disappear. Impairment of this process may lead to creation of a fistula or intramural haematoma [8]. Fistulas appearing at a later age may be a complication of myocardial infarction or occur as the consequence of a severe chest trauma [9, 10]. There is a series of iatrogenic causes leading to creation of CAF such as cardiac surgery or percutaneous interventions on coronary arteries [11[. There is also a description of fistulas complicating repeated myocardial biopsies in the process of monitoring of heart transplant rejection [13].

More than a half of fistulas remain asymptomatic and they are found accidentally usually at coronary angiography performed for other reasons. Clinical symptoms of CAF are not characteristic. Patients complain of chest pain, exertional dyspnoea, and easy fatigue. These symptoms are caused by the steal phenomenon leading to myocardial ischaemia [3]. A search for coronary fistulas should be attempted in the case of unexplained systolic-diastolic bruit on heart auscultation, especially when it is located along the left sternum margin. However, this sign was not present in our patient [14]. Fistulas may also lead to atrial fibrillation.

Studies performed by Liberthson *et al.* [15] showed that age of the patient is a deciding factor influencing the clinical manifestation and the frequency of complications caused by CAF. The authors analysed a group of 187 patients with CAF. There was a clear age difference in the presented group: patients after 20 years of age more frequently presented with clinical symptoms and complications including heart failure. Based on this research the authors suggested early surgical treatment of this malformation.

Coronary artery fistulas may be visualized using many techniques. Transthoracic echocardiography (TTE), which is a first line method, does not always visualize the fistula, as was the case in our patient [9]. The reason for low sensitivity of TTE in the detection of CAF is the small acoustic window. Transoesophageal echocardiography has advantages over TTE, because it allows precise visualization of CAF: the artery of origin, the course of the fistula and the place of drainage [16, 17]. Coronary artery fistulas may also be adequately visualized by means of computed tomography or magnetic resonance imaging [18, 19]. Coronary angiography remains a traditional method for the detection of CAF [16].

Although generally asymptomatic coronary fistulas may be complicated by infectious endocarditis, myocardial infarction, heart failure, pulmonary hypertension or coronary artery aneurysms (CAA) [16]. In extreme cases rupture of the aneurysm may cause cardiac tamponade or even sudden cardiac death if the aneurysm is large [20]

Management of coronary artery fistulas was included in the AHA/ACC guidelines from 2008 [21]. Closure of the fistula is indicated in the case of large fistulas despite the presence of clinical symptoms (class of indication I, strength of evidence C). In the case of small or moderate fistulas closure of the fistula is indicated only if it leads to ischaemia, arrhythmia or systolic/diastolic myocardial

dysfunction (I/C). Invasive treatment is not recommended in patients with small asymptomatic fistulas (III/C). This group of patients should undergo echocardiographic examinations every 3-5 years to assess any progression of pathological changes caused by the disease (IIa/C). It should be noted that these guidelines have low strength of evidence and are not very detailed. This is related to the fact that CAF is a rare disorder and therefore there are no studies on large groups of patients.

There are two methods of closing the fistula if that is necessary. The first one is percutaneous intervention using classical metal coils: Amplatzer duct occluder or Rashkind double umbrella device. The second possibility is classic cardiac surgery using extracorporeal circulation and both extracardiac and intracardiac closure of the fistula. Efficacy of both methods is similar, but percutaneous intervention is much safer.

Due to the lack of the patient's consent for cardiac surgery and stable clinical course of the disease over 2 years of follow-up with no signs of haemodynamic significance of the fistula in the presented case we decided to treat the patient conservatively with regular clinical and echocardiographic follow-up.

References

1. Lowe JE, Oldham HN Jr, Sabiston DC Jr. Surgical management of congenital coronary artery fistulas. Ann Surg 1981; 194: 373-380.
2. Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. Cathet Cardiovasc Diagn 1995; 35: 116-120.
3. Angelini P. Coronary artery anomalies-current clinical issues. Definition, classification, incidence, clinical relevance and treatment guidelines. Tex Heart Inst J 2002; 29: 271-278.
4. Demirkilic U, Ozal E, Bingol H, et al. Surgical treatment of coronary artery fistulas: 15 years' experience. Asian Cardiovasc Thorac Ann 2004; 12: 133–138.
5. Yang Y, Bartel T, Caspari G, et al. Echocardiographic detection of coronary artery fistula into the pulmonary artery. Eur J Echocardiography 2001; 2: 292-294.
6. Kassaian SE, Mahmoodian M, Salarifar M, et al. Stent-graft exclusion of multiple symptomatic coronary artery fistulae. Tex Heart Inst J 2007; 34: 199-202.
7. Krause W. Ueber den Ursprung einer akzessorischen A. coronaria aus der A. pulmonalis. Z Ratl Med 1865; 24: 225-229.
8. O'Connor WN, Cash JB, Cottrill CM, et al. Ventriculocoronary connections in hypoplastic left hearts: an autopsy microscopic study. Circulation 1982; 66: 1078-1086.
9. Radomski M, Kubica J, Sukiennik A, et al. Przetoka łącząca gałąź międzykomorową przednią lewej tętnicy wieńcowej ze światłem prawej komory: rzadkie powikłanie zawału mięśnia sercowego. Folia Cardiol 1999; 6: 399-403.
10. Jones RC, Jahnke EJ. Coronary artery-atrioventricular fistula and ventricular septal defect due to penetrating wound of the heart. Circulation 1965; 32: 995-1000.
11. Tami LF. Coronary artery-right ventricular fistula after coronary artery bypass grafting. Clin Cardiol 1993; 16: 155-157.
12. Sandhu JS, Uretsky BF, Zerbe TR, et al. Coronary artery fistula in the heart transplant patent. A potential complication of endomyocardial biopsy. Circulation 1989; 79: 350-356.
13. Akashi H, Tayama E, Tayama K, et al. Rupture of an aneurysm resulting from a coronary artery fistula-a case report. Circ J 2003; 67: 55-553.
14. Sherwood MC, Rockenmacher S, Colan SD, Geva T. Prognostic significance of clinically silent coronary artery fistulas. Am J Cardiol 1999; 83: 407-411.
15. Liberthson RR, Sagar K, Berkoben JP, et al. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. Circulation 1979; 59: 849-854.
16. Yang Y, Bartel T, Caspari G, et al. Echocardiographic detection of coronary artery fistula into the pulmonary artery. Eur J Echocardiogr 2001; 2: 292-294.
17. Abdelmoneim SS, Mookadam F, Moustafa SE, Holmes DR. Coronary artery fistula with anomalous coronary artery origin: a case report. J Am Soc Echocardiogr 2007; 20: 333.e1-4.
18. Versaci F, Del Giudice C, Sperandio M, et al. A case of coronary artery fistula visualized by 64-slice multidetector CT. Nat Clin Pract Cardiovasc Med 2009; 6: 57- 60.
19. Said SA, Hofman MB, Beek AM, et al. Feasibility of cardiovascular magnetic resonance of angiographically diagnosed congenital solitary coronary artery fistulas in adults. J Cardiovasc Magn Reson 2007; 9: 575-583.
20. Akashi H, Tayama E, Tayama K, et al. Rupture of an aneurysm resulting from a coronary artery fistula: a case report. Circ J 2003; 67: 551-553.
21. Warnes CA, Williams RG, Bashore TM, et al. American College of Cardiology; American Heart Association Task Force on Practice-ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 2008; 52: 1-121.